Juxtaductal Aortic Atresia Masquerading as Coarctation

Obed Onuzo, Michael Rigby, and Andrew Redington
Royal Brompton National Heart and Lung Hospital, London, UK

SUMMARY. Two cases of juxtaductal aortic atresia diagnosed as coarctation on clinical and Doppler echocardiographic grounds are presented. The misleading nature of the Doppler flow velocity characteristics in this condition is discussed and raises questions as to the source of these flow velocities in coarctation.

KEY WORDS: Juxtaductal aortic atresia — Coarctation — Doppler ultrasound

There have been scattered reports of interruption of the aortic arch without communication between the great arteries or any ventricular septal defect [7, 8]. In most cases there was a lack of direct continuity between the ascending and descending aorta (true interruption) [4, 11], while in others there was mural continuity in the presence of luminal atresia [3, 5]. In this report we describe two additional cases of this latter type, both of which were incorrectly diagnosed as having severe coarctation of the aorta on the basis of the aortic Doppler flow characteristics.

Case Reports

Case 1

A 4½-year-old male child presented with an asymptomatic heart murmur. On examination he was pink and was not breathless. Pulse was 90/min and blood pressure was 140/90 in the right arm. The pulses were equal in both upper limbs but his femoral pulse was weak and delayed. On auscultation the heart sounds were normal and there was a grade 5/6 ejection systolic murmur maximal at the mid-left sternal edge. No murmurs were heard over the back.

The electrocardiogram (ECG) and chest radiograph were normal. Cross-sectional echocardiography showed mild left ventricular hypertrophy. The aortic valve was trileaflet with an eccentric commissure between the left coronary and noncoronary leaflets. The Doppler velocity across the valve was 2 m/s. The aortic arch views appeared to show severe coarctation. Furthermore, the peak instantaneous velocity on stand-alone continuous wave Doppler was only 1.3 m/s in the descending aorta. There was an associated long diastolic pressure decay time with a time to half peak diastolic gradient of 115 m (Fig. 1).

Cardiac catheterization was performed with a view to balloon dilatation of the aortic coarctation. The aortic arch could not be entered from the descending aorta, and the aortogram through the right brachial artery showed a blind-ending aortic arch, just distal to the left subclavian artery.

At surgery, the aortic arch was in continuity with the descending aorta but completely atretic. The atretic segment was excised with end-to-end anastomosis. Microscopic examination confirmed complete atresia with a central dimple on both aspects of the segment. Postoperative course was complicated by mild systemic hypertension (blood pressure 160/90) which was treated with propranolol.

Case 2

A 5-month-old male patient was admitted to his local hospital with heart failure. A diagnosis of myocarditis was made and he was treated with digoxin. His left ventricular function, which was initially poor, improved and treatment was discontinued after 2 years. He continued to have a murmur which on echocardiography was attributed to a muscle band in the left ventricle.

Although he remained asymptomatic, weak femoral pulses were noticed at 5 years of age. Therefore, he was referred for further investigations.

On examination, the upper limb pulses were normal but the femoral pulses were weak with radiofemoral delay. Blood pressure was 150/90 in both arms and 100 systolic in the left leg. A grade 2/6 ejection systolic murmur was heard at the lower left sternal edge. There were no other significant abnormalities. The chest radiograph showed a normal-sized heart and a clear indentation ("figure of 3" sign) of the descending aorta but no rib notching. The ECG showed an atrial rhythm with a P-wave axis of −45 and a mean frontal QRS axis of +70. There was left ventricular hypertrophy on voltage criteria, but no T-wave abnormalities.

Cross-sectional echocardiography confirmed normal intracardiac anatomy. The arch views again suggested coarctation,

Address offprint requests to: Dr. Andrew Redington, Royal Brompton National Heart and Lung Hospital, Sydney Street, London SW3 6NP, UK.
but no systolic jet could be detected with color-flow Doppler. However, there was a peak instantaneous systolic velocity measured with continuous wave Doppler of 4.2 m/s in the descending aorta. The diastolic velocity decay was markedly prolonged, such that the velocity at end-diastole was still greater than half of the peak.

Angiography showed a blind-ending descending aorta (Fig. 2) and the ascending aorta could not be entered. The left subclavian artery was not outlined from the descending aorta.

At surgery, there was an atretic segment between the left subclavian artery and the descending aorta. Resection with end-to-end anastomosis was performed.

The postoperative course was uneventful.

Discussion

The first case of interrupted aortic arch with normal intracardiac anatomy was described in an autopsy series by Evans in a 32-year-old man in 1933 [2]. In 1964, Pillsbury et al. described the first live case in a female aged 16 years [9]. Both of these cases, and most others described in the literature, had true interruption of the aorta with a lack of continuity between ascending and descending aorta. In our cases, while blood flow from ascending to descending aorta was "interrupted," the anatomy was more typical of severe coarctation with probably acquired atresia.

The use of Doppler echocardiography in the diagnosis of coarctation has been well described [10, 12] and its deficiencies noted [6]. More recently, data from our unit, published by Carvalho et al., described the criteria for assessment of severity of coarctation using a combination of peak systolic gradient and time to half-diastolic gradient [1]. Either a peak systolic gradient or 40 mmHg or more or a time to half-diastolic gradient equal to or greater than 100 ms was significant in discriminating between lesions requiring intervention and those that did not.

The overall shape of the Doppler spectral of the two patients described was typical of severe coarctation, and in both patients fulfilled the criteria for intervention. The Doppler data were misleading, however, as there was atresia of the aortic arch in both. Almost certainly the descending aortic flow velocities that were recorded reflected flow in or from collaterals. These characteristics are therefore potentially misleading, particularly if balloon dilatation is considered: both of our patients had undergone cardiac catheterization unnecessarily.

It is difficult to see how these errors can be avoided. Color-flow mapping may be suggestive of atresia but it is of poor specificity, particularly in the older patient. Magnetic resonance imaging may be useful, particularly if velocity mapping is performed, but it cannot be applied to all patients.

In summary, acquired juxtaductal atresia of the aortic arch is a rare but important part of the spectrum of coarctation of the aorta. Doppler echocardiography may be misleading and will show diastolic flow velocities similar to those seen in many patients with severe coarctation. The diastolic